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Case report

Giant intradural cervical spine arteriovenous malformations – A case and review of literature

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ARTICLE INFO

Article history:
Received 8 July 2017
Accepted 27 February 2018
Available online xxx

Keywords:
Cervical spine
Arteriovenous malformations
Angiography
Embolization
Endovascular

ABSTRACT

Introduction: Spinal arteriovenous malformations (SAVMs) are very rare and can be very challenging to treat since none of the therapeutic options does provide a definitive cure to these lesions. We believe that incorporation of intraoperative angiography during surgery in a hybrid theatre can help achieve a better cure.

Case presentation: We present a 45 years old woman with three (3) years history of weakness and ten (10) days' history of acute pain in right upper extremity. Magnetic resonance angiography (MRA) of the cervical segment of spinal cord revealed tortuous vascular masses from foramen magnum to the inferior margin of fourth cervical (C4) vertebral. Spinal digital subtracting angiography (DSA) confirmed vascular malformation at the cervical segment of the spinal cord with their origin from bilateral posterior spinal arteries. She was successfully operated on with the aid of intraoperative angiography without any neurological deficient.

Conclusion: Spinal angiography is the gold standard for all-inclusive assessment of SAVMs. Surgery and endovascular techniques equally have key therapeutic valves in treatment of SAVMs but a combination of the two gives a more accurate and reliable cure to this disorder.

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Abbreviations: AV, arteriovenous; AVF, arteriovenous fistulas; AVMs, arteriovenous malformations; C, cervical vertebral; CT-scan, computer tomographic scan; L, lumber vertebral; DSA, digital subtracting angiography; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; SAH, subarachnoid haemorrhage; SAVM, spinal arteriovenous malformations; SDAVF, spinal dural arteriovenous fistula; T, thoracic vertebral.

https://doi.org/10.1016/j.pjnns.2018.02.009
0028-3843/

1. Introduction

Spinal arteriovenous malformations (SAVM) are rare [1–4] and in conjunction with other spinal cord vascular disorders constitutes about 3–4%, and can be related to significant neurological disabilities and even death if not detected and treated early [1,5]. Cervical SAVMs are even more rarer and so far, only a hand full of literature have reported arteriovenous malformations (AVMs) in this location. Cervical SAVMs are very challenging to detect since their clinical course imitate a diverse range of neurological conditions [6]. They can be grouped into high-flow and slow-flow base on their pathophysiology. While high-flow lesions may lead to ischaemia or haemorrhage, slow flow on the other hand may lead to venous congestion, compression of spinal cord and ischaemia [7]. Based on their location in the cord, angioarchitecture, intradural or extradural and/or presence or absence of arteriovenous (AV) shunts, SAVMs can be categorized into four (4) types [7,8]. SAVMs are curable disorders [9] and recent improvements in neuro-imaging and endovascular techniques outlining the angioarchitecture of spinal vascular anatomy have led to enhanced treatment of this disorders [10]. We report a case of giant intradural cervical SAVM which successfully treated by incorporates both open surgery and intraoperative angiography without any neurological deficient.

2. Case report

We present a 45 years old woman with three (3) years history of weakness and ten (10) days' history of acute pain in right upper extremity. She was apparently diagnosed 3 years ago with spinal vascular malformations in a local hospital with no treatment. She has been visiting the local hospital for pain relievers intermittently until ten (10) days prior to presentation at your facility when her condition aggravated. The weakness and pain at right upper limb radiated to the neck with persistent needle-like pain (Paresthesia). She however has no numbness as well as incontinence. Her bladder and bowel habits have not changed. She was put on Gabapentin capsules for the pain relieve. Past medical and surgical history was unremarkable.

General physical examination did not yield much. All the systems were grossly normal. Neurological examination revealed intact cranial nerves. On the upper limbs, the right arm had corresponding weakness of 3/5 proximal strength and 4/5 strength in his distal muscles while the left arm had corresponding weakness of 4/5 proximal strength and 4/5 strength in his distal muscles. Corresponding dermatomes were normal. She had brisk and bizarre sensations to pin prick, cold and hot stimuli on the upper arms but normal in lower limbs. Proprioception and vibratory sense were markedly reduced in the upper arm but normal in the lower limbs. Abdominal reflexes were absent and rectal tone intact. She low limbs however had bilateral corresponding 5/5 proximal strength and 5/5 strength in his distal muscles. She could walk about with any form of support. All reflexes were present and normal. Routine laboratory and other ancillary investigation were normal.

Magnetic resonance angiography (MRA) of the cervical segment of spinal cord revealed tortuous vascular masses from foramen magnum to the inferior margin of C4 vertebral. The lesions are significantly heterogeneously enhanced. The spinal cord significantly compressed the masses, so parts of the spinal cord are not visible. The vertebra disc from C2–C7 is somehow collapsed with posterior protrusion of the edges of vertebral bodies resulting in the relative compression of the dura sac. On T2-MRI, cervical intervertebral discs have iso to hyper-intensity. There are no para-spinal soft tissue swellings. A working diagnosis of intra-spinal vascular malformation from foramen magnum to the inferior margin of C4 vertebral was made (Fig. 1A and B). Spinal digital subtracting angiography (DSA) was done at the local hospital and repeated at our facility showed vascular malformation at the cervical segment of the spinal cord with their origin from bilateral posterior spinal arteries (Fig. 2A–D). After educating and counselling the patient as well as her relatives, operation was scheduled.

The operation was done in our hybrid surgical theatre that incorporates both open surgery and intraoperative angiography. The aim of our surgery was to excise this giant vascular malformation in cervical vertebral canal with selected angioplasty, decompression of vertebral canal and spinal nerve root as well as fixation of fused posterior C2–C6. The patient was put on the prone position with the head rested on the doughnut head support after general anaesthesia. The skin marking was down from mid occipital hairline to about C6 vertebral. The endovascular surgeon also draped the left inguinal region and cannulated the left femora artery. Draping of the neck was done after securing the artery and the cannula inserted.

The skin incision was made and the subcutaneous, intramuscular, along the C2–C7 was dissected carefully. The para-spinal muscles were separated from the spinous processes at both sides followed by exposition of the lower part of the occiput at the foramen magnum, the atlas arch C2–C7 removed and laminectomy done. The dura was opened and we saw huge tortuous intradural vascular malformation that were more localized to the left side and extended from C1–C5. These abnormal vascular masses were originating from both posterior spinal arteries. They cause enlargement of the spinal canal with extensive adhesions and compressing of nerve roots as well as deformation (Fig. 3A).

Intraoperative angiography was used to identify the feeding arteries intra-operatively and they were resected with no complication. The microscopy was used throughout the operation. After total resection of the lesion, intraoperative angiography was done again which confirm total resection of the lesions. Intraoperative images also show total resection (Fig. 3B and C). After securing total hemostasis within spinal cord, the dura was repaired with artificial dura mater and posterior C2–C4 fixation done with titanium screws. A drainage tube was inserted and suturing of muscle, fascia, as well as subcutaneous tissue and skin. (The resected SAVM is as shown in Fig. 3D.) The patient recovered very quickly with no neurological deficits and the pain at the left arm was improved. Postoperative MRI was done and this revealed no residual SAVM (Fig. 4A and B). She was put on a rigid cervical colour for six (6) months. She was discharged home two (2) weeks after the operation and schedule outpatient visits arranged every three (3) months.

Please cite this article in press as: Richard SA, et al. Giant intradural cervical spine Q1arteriovenous malformations – A case and review of literature. Neurol Neurochir Pol (2018), https://doi.org/10.1016/j.pjnns.2018.02.009